

Her most recent chest x-ray film (December 20, 1983) was unchanged from the previous one (Figure 2).

Discussion

In summary, this patient presented as a 44-year-old woman with a ten-year history of multifocal eosinophilic granuloma. There was initial involvement of the lungs with subsequent development of pituitary and bony lesions. The age of the patient and the progressive course of the disease are uncommon in the presentation of eosinophilic granuloma.

It is interesting that our patient had a pneumothorax. The association of diabetes insipidus and pneumothorax in multifocal eosinophilic granuloma has previously been noted in the literature. Lewis¹ reviewed the clinical features of 75 patients diagnosed as having eosinophilic granuloma by lung biopsy. Six of seven patients with diabetes insipidus also had pneumothorax. However, Friedman and co-workers,² in their discussion of 60 cases with pulmonary involvement, noted that of five patients who had diabetes insipidus, none had pneumothorax. The authors also pointed out that further systemic involvement was unusual (none in their series) following presentation with primary pulmonary lesions.

Pulmonary disease in our patient appeared to be characteristic of that in eosinophilic granuloma. Evaluation of pulmonary function showed a pattern of decreased diffusion capacity and restricted lung volumes. There was not reduction in air flow.

Eosinophilic granuloma of the lung in our patient was treated initially with a regimen of prednisone, 60 mg a day, with objective and subjective improvement. However, low-dose therapy with 10 mg of prednisone on alternate days seemed to give only subjective relief of the symptoms with continuous progression of pulmonary disease. It is possible that in the absence of the steroid treatment, a more rapid course of the disease would have taken place.

A similar statement could be made as far as treatment with vinblastine in our patient. Over the period of time that vinblastine was administered there was no deterioration as shown by pulmonary function test results. However, neither was there any improvement. Whether this pattern was part of the natural course of the disease in this particular patient is unknown.

Evidence of hypothalamic-pituitary axis involvement became apparent in our patient about a year and a half after the initial presentation. At that time dysfunction of the posterior pituitary was evident based on the results of the fluid deprivation test. Diabetes insipidus is the most common endocrinopathy of multifocal eosinophilic granuloma and is well documented in the literature.³⁻⁵ It has also been pointed out in the literature that primary lesions of the anterior pituitary are quite uncommon and any abnormality of function can be ascribed to lesions in the hypothalamus.⁶⁻⁸

There was an increase in the basal level of prolactin and a decrease in both the basal gonadotropin levels and their response to gonadotropin-releasing hormone stimulation with the development of secondary amenorrhea and galactorrhea. These abnormalities are probably due to the pituitary stalk lesion observed by computed tomography.

REFERENCES

1. Lewis JG: Eosinophilic granuloma and its variants with special reference to lung involvement—A report of 12 patients. *Q J Med* 1964 Jul; 33:337-359

2. Friedman PJ, Liebow AA, Sokoloff J: Eosinophilic granuloma of lung—Clinical aspects of primary pulmonary histiocytosis in the adult. *Medicine (Baltimore)* 1981 Nov; 60:385-396

3. Lieberman PH, Jones CR, Dargeon HWK, et al: A reappraisal of eosinophilic granuloma of bone, Hand-Schüller-Christian syndrome and Letterer-Siwe syndrome. *Medicine (Baltimore)* 1969 Sep; 48:375-400

4. Braunstein GD, Kohler PO: Pituitary function in Hand-Schüller-Christian disease—Evidence for deficient growth-hormone release in patients with short stature. *N Engl J Med* 1972 Jun 8; 286:1225-1229

5. Kaufman A, Bukberg PR, Werlin S, et al: Multifocal eosinophilic granuloma ('Hand-Schüller-Christian disease'). *Am J Med* 1976 Apr; 60:541-548

6. Rothman JG, Snyder PJ, Utiger RD: Hypothalamic endocrinopathy in Hand-Schüller-Christian disease. *Ann Intern Med* 1978 Apr; 88:512-513

7. Goodman RH, Post KD, Molitch ME, et al: Eosinophilic granuloma mimicking a pituitary tumor. *Neurosurgery* 1979 Dec; 5:723-725

8. Gates RB, Friesen H, Samaan NA: Inappropriate lactation and amenorrhoea: Pathological and diagnostic considerations. *Acta Endocrinol (Kbh)* 1973 Jan; 72:101-114

Kaposi's Sarcoma Presenting as Homogeneous Pulmonary Infiltrates in a Patient With Acquired Immunodeficiency Syndrome

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AGGRESSIVE KAPOSI'S SARCOMA is a well-described occurrence in patients with acquired immunodeficiency syndrome.¹ Even when the disease is widely disseminated, however, the chest radiograph is usually normal.² Although Kaposi's sarcoma has been reported in the lung,³ we report the first case of Kaposi's sarcoma in which the initial clinical manifestation was a rapidly progressive, homogeneous pulmonary infiltrate. This case stresses the importance of considering Kaposi's sarcoma in the differential diagnosis of pulmonary infiltrates, even in the absence of other visceral or dermatologic manifestations.

Report of a Case

The patient, a 30-year-old homosexual man, was admitted to the University of California Irvine Medical Center with complaints of malaise, weight loss, alopecia and bright red blood per rectum for two months. A test for hepatitis B surface antigen was positive. The patient had herpes proctitis (confirmed by clinical diagnosis and by culture results) and *Giardia lamblia* enteritis. A chest roentgenogram showed infiltrates involving the left upper and lower lobes and the right midlung field. A diagnosis of *Pneumocystis carinii* pneumonia was made by bronchoscopic biopsy and trimethoprim-sulfamethoxazole therapy was initiated. Erythema multiforme developed, but resolved when the trimethoprim-sulfamethoxazole regimen was discontinued. Pentami-

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dine therapy was begun and continued for a total of 21 days with subsequent pronounced improvement as shown by the chest roentgenogram and by the patient's physical condition. Results of immunologic studies were consistent with a diagnosis of acquired immunodeficiency syndrome. The patient was discharged.

Three weeks later he returned to hospital with fever, chills and cough productive of yellow sputum. At this time he appeared cachectic, but was afebrile. A careful physical examination showed no skin lesions and only a few shotty, small, anterior cervical lymph nodes. Lungs were clear to auscultation; the abdomen was soft, nontender and without hepatosplenomegaly. A rectal examination showed scarring in areas of previous proctitis. The stool was brown and a stool specimen was positive for trace amounts of occult blood. A chest roentgenogram showed a residual patchy infiltrate involving primarily the left upper lobe.

The patient's hospital course was complicated by low-grade fevers with occasional fever spikes. All cultures were negative. Sputum smears and bronchoscopy were nondiagnostic. A bone marrow specimen was considered normal. At six weeks, blood, sputum and marrow cultures became positive for *Mycobacterium intracellulare avium* and antimycobacterial therapy was initiated. Rectal bleeding developed and a sigmoidoscopic examination showed a rectal fissure and old scarring. Cultures were negative for herpes. The patient's mental state deteriorated gradually although his physical state was stable until the 58th hospital day, when his temperature abruptly reached 40°C (104°F) and he had diffuse rhonchi, increased sputum and mild respiratory distress. A chest roentgenogram taken at this time showed new, diffuse alveolar and interstitial infiltrates involving both lung fields, radiographically a more extensive but somewhat similar picture to the earlier findings of *Pneumocystis* (Figure 1). Consideration was given to simply initiating pentamidine therapy. However, specimens taken by bronchoscopy and stained showed no *Pneumocystis* or other pathogens. Results of open-lung biopsy showed only Kaposi's sarcoma. Eight days later, the patient died.

Autopsy showed Kaposi's sarcoma involving the rectum and retroperitoneal and mediastinal lymph nodes with diffuse involvement of the lungs (Figures 2 and 3). Periodic acid-Schiff stains, Giemsa stains, hematoxylin and eosin stains, methenamine silver stains, routine cultures, fungal cultures and mycobacterial cultures were done. The results of all studies were negative except that at eight weeks the lung autopsy specimen grew a few colonies of *M avium*.

Discussion

Kaposi's sarcoma in homosexual patients with acquired immunodeficiency syndrome is extremely aggressive⁴ and can involve virtually any organ system.¹ The lung, however, is rarely involved.² Reports of lung involvement are nearly always of an indolent reticular-nodular infiltrate noted in the presence of sarcoma of the skin or lymph nodes.⁵ Kornfeld and Axelrod have reported a case of acquired immunodeficiency syndrome with nodular, interstitial infiltrates from Kaposi's sarcoma: this patient had no other obvious sarcoma elsewhere.³ They described the case of a patient with a subacute course in whom *P carinii* pneumonia developed after several months and who died thereafter of respiratory failure.

The composite, therefore, of a patient who has acquired immunodeficiency syndrome with pulmonary Kaposi's sarcoma is a homosexual man who has obvious sarcoma in the skin, lymph nodes or other viscera, and in whom hilar adenopathy then develops with nodular or linear infiltrates. These patients frequently have or will go on to have concomi-

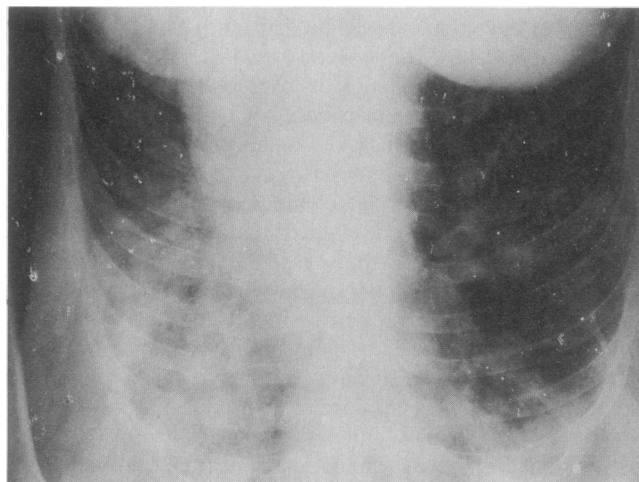


Figure 1.—Roentgenogram of patient's chest taken just before open-lung biopsy, showing new diffuse alveolar and interstitial infiltrates.

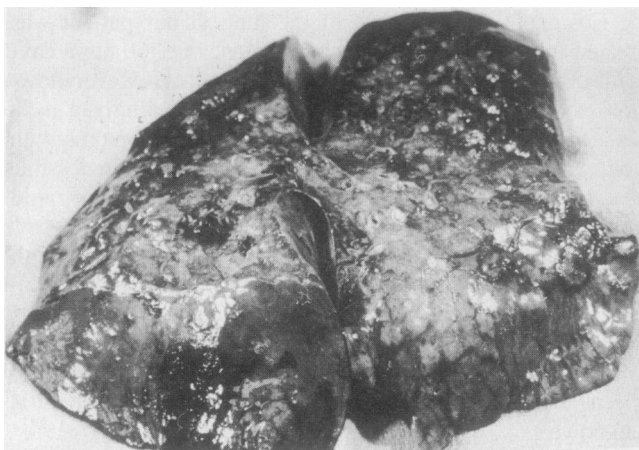


Figure 2.—Gross lung specimen taken at autopsy.

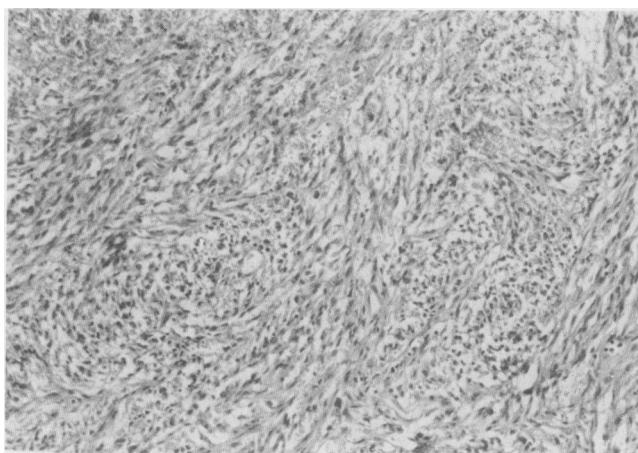


Figure 3.—Tumor growing into alveolar walls (× 120).

CASE REPORTS

tant pulmonary infection with any of a variety of agents including *P carinii*, cytomegalovirus, *Cryptococcus*, *Toxoplasma*, *Aspergillus*, *Candida*, *Legionella*, *Histoplasma* and *Pettriellidium boydii*.^{1,5}

Our patient is unique in several respects. Most important, this is the first report of pulmonary Kaposi's sarcoma presenting as an aggressive, alveolar infiltrate progressing rapidly to respiratory failure and death. Extensive pathologic evaluation and many cultures were done. The only evidence of other pathologic features was the growth of a few colonies of *M avium*. We feel strongly that the infiltrate was due to Kaposi's sarcoma and not *M avium* for the following reasons: The patient was being treated with five antimycobacterial drugs for *M avium*, which had previously been identified in the central nervous system, and the infiltrate occurred very rapidly; the acid-fast bacilli stains were entirely negative on both biopsy and autopsy specimens, and the biopsy specimens showed extensive tumor invasion of the parenchyma and possible disruption of the alveolar wall.

This case also serves to remind clinicians that Kaposi's sarcoma can present first in the lung. At autopsy some retroperitoneal and mediastinal nodal involvement was found that was not apparent clinically. In addition, the rectal involvement was attributed to scarred herpetic proctitis and therefore a connection with Kaposi's sarcoma was overlooked.

When rapidly progressive homogeneous infiltrates appear in patients with acquired immunodeficiency syndrome, the most common cause is *P carinii*.⁵ However, all of the organisms mentioned above must be considered. Although Kaposi's sarcoma generally causes indolent, nodular infiltrates in patients whose sarcoma is apparent in other organs, our case suggests that Kaposi's sarcoma should also be included as a possible aggressive pneumonitis. The recognition of this phenomenon is important for at least two reasons. First, because, as in our patient and in others,⁶ a clinician may consider administering inappropriate antibacterial therapy to a patient; and, second, because aggressive chemotherapeutic regimens may prove at least partially effective in the treatment of Kaposi's sarcoma.⁴

REFERENCES

1. Groopman JE: Kaposi's sarcoma and other neoplasms. In Gottlieb MS (Moderator): The acquired immunodeficiency syndrome. Ann Intern Med 1983 Aug; 99:208-220
2. Mann SG: Kaposi's sarcoma. AJR 1974 Oct; 121:793-799
3. Kornfeld H, Axelrod JL: Pulmonary presentation of Kaposi's sarcoma in a homosexual patient. Am Rev Respir Dis 1983 Feb; 127:248-249
4. Friedman-Kien AE, Laubenstein LJ, Rubinstein P, et al: Disseminated Kaposi's sarcoma in homosexual men. Ann Intern Med 1982 Jun; 96(Pt 1):693-700
5. McCauley DI, Naidich DP, Leitman BS, et al: Radiographic pattern of opportunistic lung infections and Kaposi's sarcoma in homosexual males. AJR 1982 Oct; 139:653-658
6. Brown RK, Huberman RP, Vanley G: Pulmonary features of Kaposi's sarcoma. AJR 1982 Oct; 139:659-660